中文題目：罕見以肝動脈動脈瘤破裂表現的結節性多發動脈炎-案例報告

英文題目：Polyarteritis nodosa rarely presenting as hepatic artery aneurysm rupture: a case report

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Introduction：

Polyarteritis nodosa (PAN) is a rare necrotizing vasculitis and is more frequent in men than in women. It occurs most frequently in patients in their 40s to 60s. It may involve multiple organs including kidneys, skin, peripheral nerves, striated muscle, and the gastrointestinal tract. Vasculitis may cause aneurysm and thrombosis formation. As a result, aneurysm rupture and mesenteric ischemia are two most life-threatening complications. Prompt diagnosis and appropriate intervention are important for survival. Clinical manifestation detection, imaging and pathology are crucial for diagnosis of PAN.

Case presentation：

We reported a 60-year-old man with chief complaint of body weight loss of five kilogram in the past three months, black stool passage and intermittent epigastric dull pain. Initial laboratory tests showed microcytic anemia with hypoalbuminemia. Esophagogastroduodenoscopy showed gastric healing ulcer. Computed tomography (CT) showed several hypodense lesions in bilateral renal cortex. Sputum, stool and urine tuberculosis culture, tumor marker, autoimmune profile all showed negative results.

We noticed livedo reticularis and the patient complained of right testicular pain after admission. Besides, he then developed acute onset diffuse abdominal pain on the eleventh hospitalization day.Abdominal CT showed heterogeneous density around left liver lobe with contrast media extravasation, multiple wedge shape hypoperfusion lesions in both kidneys and low-density lesion in the spleen were observed (Figure.1). Emergent celiac angiography revealed contrast medium extravasation in the branch of left hepatic artery, several abnormal contrast stains in both lobes of liver, and abutting the distal branches of bilateral hepatic arteries which suspected microaneurysms (Figure.2). He received transcatheter arterial embolization (TAE) for the internal bleeding and was transferred to Intensive Care Unit.

Under the tentative diagnosis of PAN with intra-abdominal infection, he received steroid and antibiotic treatment. However, he developed another episode of shock on seventeenth hospitalization day. Repeated abdominal CT disclosed pneumoperitoneum

and poor wall enhancement of ileum. He received emergent exploratory laparotomy and small intestine resection due to small bowel ischemic change with perforation from jejunum to terminal ileum. Pathology reported transmural ischemic necrosis with dense acute inflammatory cell infiltration and atherosclerosis in mesentery vessels. Repeated diagnostic angiography for renal artery, superior mesenteric artery (SMA), inferior mesenteric artery (IMA) and celiac trunk showed multiple microaneurysms at bilateral hepatic arteries, SMA, IMA, and bilateral renal arteries (Figure.3).

He also had massive bloody stool on fifty-second hospitalization day. CT showed contrast medium extravasation over cecum and ascending colon. Angiography showed contrast medium extravasation at branch of ileocolic artery around cecum. He received TAE again for the bleeding control. Unfortunately he finally expired due to concomitant nosocomial infection including pneumonia, intra-abdominal infection, bacteremia and fungemia.

Discussion：

PAN is defined as vasculitis with the involvement of medium sized vessels rather than smaller vessels or glomeruli as well as the absence of antineutrophil cytoplasmic antibodies (ANCA). Clinical characteristics of PAN include systemic symptoms (fatigue, weight loss, weakness, fever and arthralgias) and signs (skin lesions, hypertension, renal insufficiency, neurologic dysfunction and abdominal pain) of multisystem involvement. There is no specific serologic test for PAN. Criteria for the diagnosis of PAN has been applied with sensitivity and specificity of 82 and 87% respectively. Our patient met the criteria with significant unintentional weight loss (above 5 kg within past three months), livedo reticularis over upper limbs, weakness, diastolic blood pressure above 90 mmHg, elevated blood urea nitrogen, elevated creatinine level, and microaneurysm detected by angiography.

The natural course of untreated systemic PAN is rapidly progressive and can be fatal. Those patients with older age and gastrointestinal complication had higher mortality. Around 60 percent of PAN patients have aneurysm formation in the hepatic vessels detected by autopsy. However, rupture of hepatic artery aneurysm and ileocolic artery aneurysm is rarely reported. Our patient suffered from the rare presentation of hepatic artery aneurysm rupture and developed complication of ischemic bowel disease with perforation, ileocolic artery aneurysm rupture with massive bloody stool during hospital course. We provided TAE and surgery for invasive intervention and methylprednisolone for PAN control. As review, only 23 PAN patients with hepatic artery aneurysms were reported. All patients had CT examination and 11 patients (47%) received management

of coil embolization. Only two of these patients (8%) were diagnosed as PAN before aneurysm rupture. As a result, early detection of systemic symptoms and signs compatible with diagnostic criteria of PAN is important. Our patient had detectable lesion by CT and TAE was applied. Unfortunately, he had ischemic bowel disease during follow-up and received laparotomy with small bowel resection. As reported before, four patients (17%) died within 12 weeks after hepatic artery rupture and PAN diagnosed. Three of them died from septic shock and one died from pulmonary embolism. As described before, PAN patient with mesenteric perforation and hemorrhage had 100% mortality despite interventional efforts. Gastrointestinal involvement leads to poor prognosis. Treatment including regimen of glucocorticoids in addition to immunosuppressants are needed for PAN. Nonetheless, increased risk of infection is also the concern.

Conclusion:

PAN is a rare disease which diagnosis needs combination of clinical features, images and pathology. Early diagnosis leads to better prognosis. Complications of gastrointestinal involvement such as aneurysm rupture, mesenteric ischemia, bleeding may be fetal. Timely detection with appropriate management is crucial.

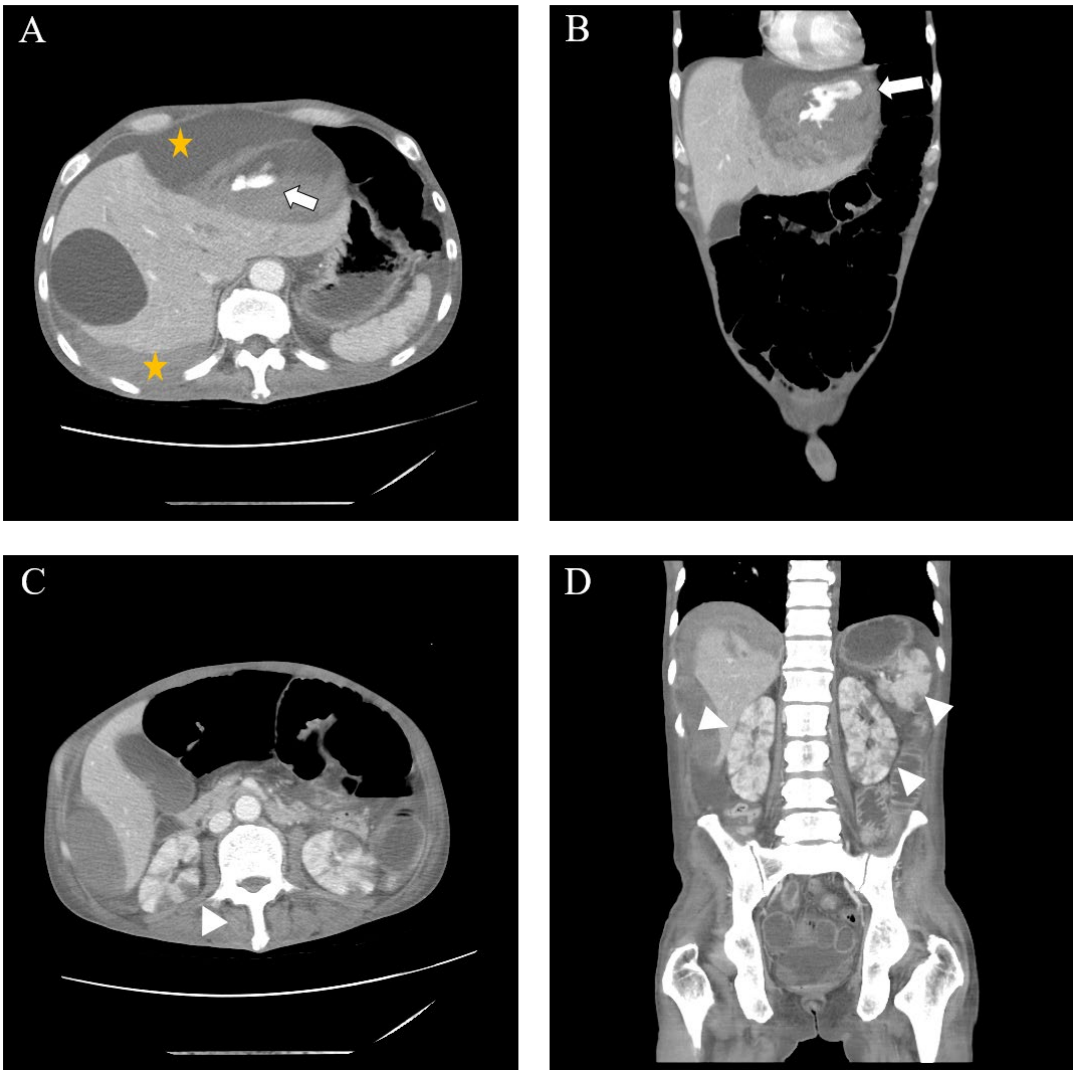

Figure 1

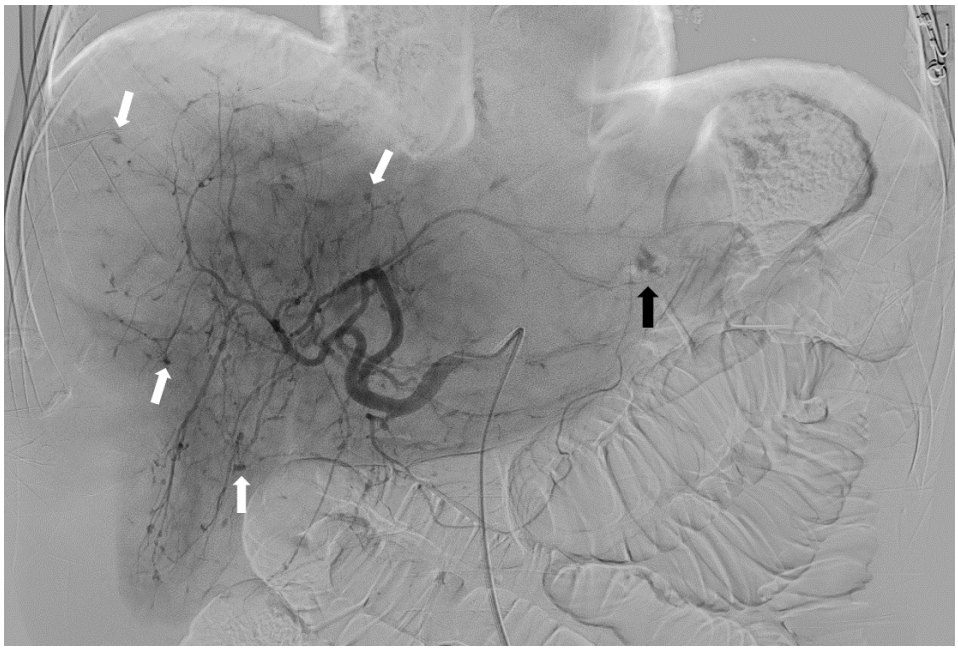

Figure 2

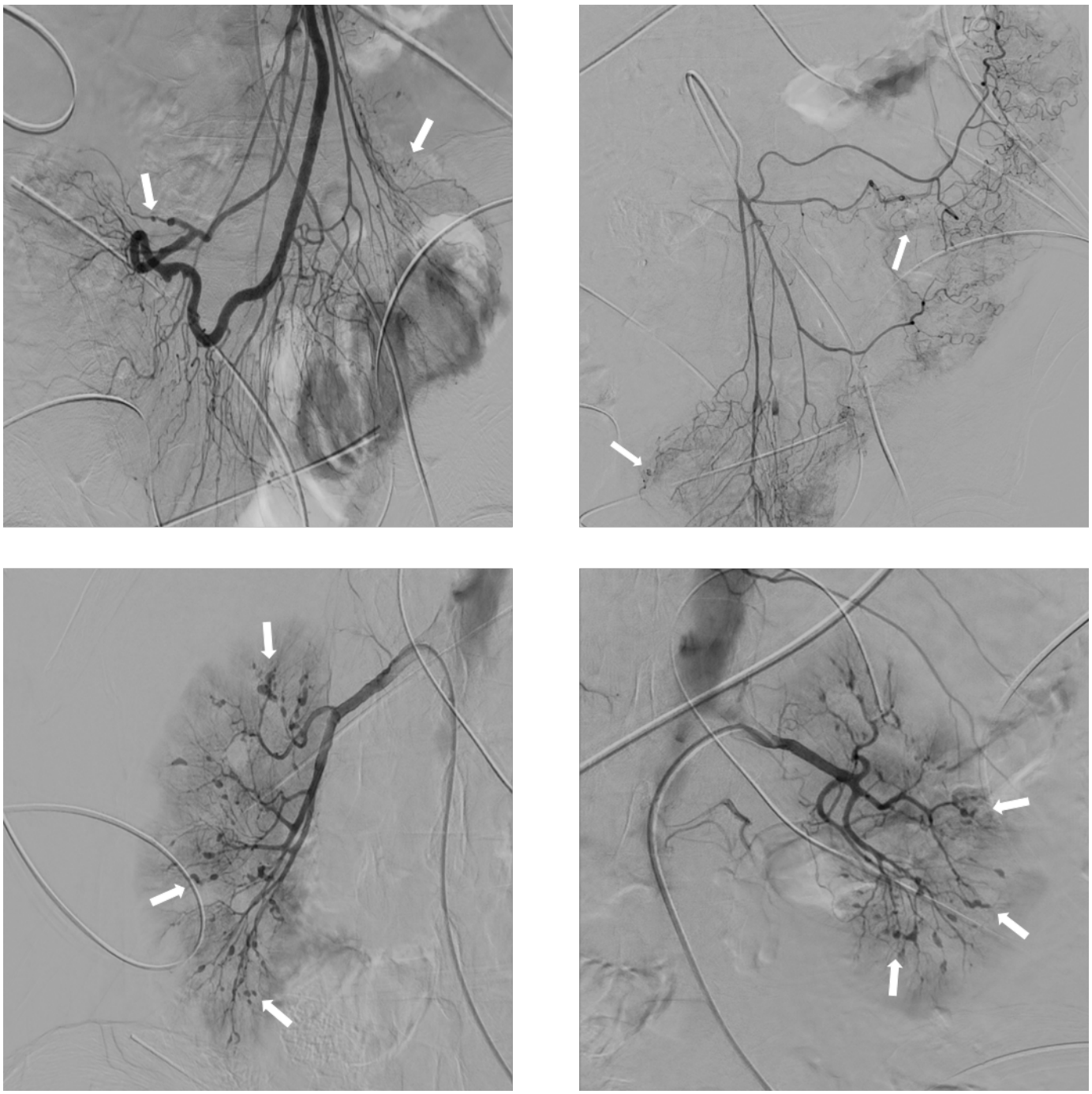

Figure 3